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Isolated Vestibular Syndrome With "Double-Panda" Sign in CNS Lymphoma

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Dear Editor,

Central nervous system (CNS) lymphoma is a rare type of non-Hodgkin lymphoma in which malignant cells from lymphoid tissue form in the brain and spinal cord (primary) or spread from other parts of the body to the brain and spinal cord (secondary).[1] CNS lymphoma can manifest with various neurological symptoms depending on its location.[2] A characteristic imaging finding of primary CNS lymphoma (PCNSL) is a computed tomography (CT) hyperdense enhanced supratentorial mass that is hypointense in T1-weighted magnetic resonance imaging (MRI) and iso- to hypointense in T2-weighted MRI, with vivid homogeneous enhancement and restricted diffusion.[1-3] We describe distinctive neuro-otological findings and the typical "double-panda" sign on brain MRI in a patient with CNS lymphoma.

A 65-year-old male presented with acute dizziness with a 3-day history. Oculography showed horizontal gaze-evoked nystagmus (GEN) during bilateral gaze, and impaired horizontal smooth pursuit (Supplementary Fig. 1A in the online-only Data Supplement). Saccades were normal. Video head impulse tests (HITs) revealed decreased vestibulo-ocular reflex (VOR) gains of both horizontal and posterior semicircular canals (Supplementary Fig. 1B in the online-only Data Supplement). Ocular vestibular evoked myogenic potentials (VEMPs) were decreased during right-ear stimulation, while cervical VEMPs were symmetric (Supplementary Fig. 1C and D in the online-only Data Supplement). The levels of serum ceruloplasmin and 24-hour urine copper were normal, and Kayser-Fleischer rings were not observed. Brain MRI showed the characteristic "double-panda" sign with additional increased T2-weighted signal intensities along the bilateral ventricular walls, thalamus, hypothalamus, mammillary body, optic tract, dorsal pons, and upper medulla, and contrast enhancement in the ependyma (Fig. 1A-C). There was slight improvement of symptoms after treatment with steroid. One month later the patient was readmitted due to a sudden decrease in mentality. Brain CT showed obstructive hydrocephalus (Fig. 1F), and an external ventricular drain was placed. The endoscopic biopsy confirmed diffuse large-B-cell lymphoma (Supplementary Fig. 2 in the online-only Data Supplement).

To the best of our knowledge, the present patient is unique since he showed the typical "double-panda" sign on brain MRI due to CNS lymphoma, which is traditionally considered to be characteristic of Wilson's disease.[4] The midbrain face of the "giant-panda" sign comprises normal-intensity red nuclei (eyes) and lateral portions of substantia nigra pars reticulata (ears), with high signal intensity in the tegmentum and hypointensity in the superior colliculus (mouth) (Fig. 1D). The pons face of the "miniature-panda" sign consists of relative hypointensity of the central tegmental tracts (eyes) contrasting with the hyperintensity of the aqueduct opening into the fourth ventricle (nose and mouth) (Fig. 1E).[4] Combining these features produces the "double-panda" sign. Our case indicates that CNS lymphoma should be considered in the differential diagnosis of this rare imaging finding if there is no clear evidence of Wilson's disease.

CNS lymphoma in our patient was supported by the lack of evidence for toxic or metabolic

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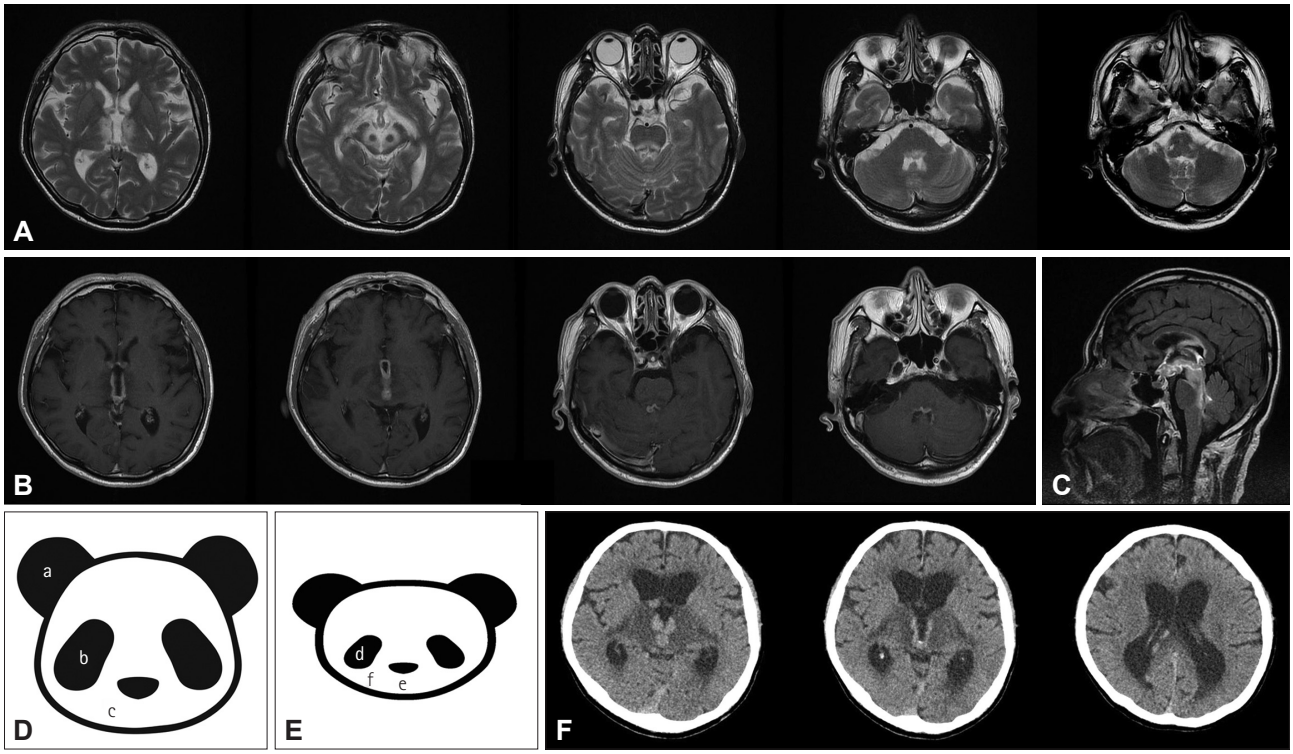

Fig. 1. Magnetic resonance imaging findings. Axial T2-weighted (A), axial gadolinium-enhanced T1-weighted (B), and sagittal T2-weighted (C) images of the patient show the "double-panda" sign with high signal intensities in the ventricular walls, thalamus, hypothalamus, dorsal pons, and upper medulla, and enhanced lesions along the ventricular walls. D and E: Schematic of the "double-panda" sign. D: The midbrain face of the "giant-panda" sign: a. substantia nigra pars reticulata (ears), b. red nucleus (eyes), and c. superior colliculus (mouth). E: Schematic of the "miniature-panda" sign: d. central tegmental tract (eyes), e. aqueduct opening into the fourth ventricle (nose and mouth), and f. superior cerebellar peduncle (cheeks). F: Brain computed tomography reveals obstructive hydrocephalus with hyperdense lesions along the ventricular walls.

disorders, the partial response to steroid treatment, and the additional involvement of the periventricular areas on brain MRI. PCNSL mostly involves brain parenchyma as a primary site of CNS involvement, preferentially with single or multiple enhanced lesions in the periventricular and superficial brain.[1,3,5] A strongly enhanced lesion with diffusion restriction adjacent to the subarachnoid space and a lesion crossing the corpus callosum are characteristic MRI findings of PCNSL.[1]

Remarkably, our patient presented with isolated vestibular syndrome with characteristic findings involving the vestibular nucleus, which has rarely been described in CNS lymphoma. Recently, a patient with solitary PCNSL also developed acute dorsal medullary syndrome with abnormal vestibular findings affecting the vestibular nucleus.[5] Lesions restricted to the vestibular nuclei produce the signs of both unilateral peripheral vestibulopathy (spontaneous nystagmus, caloric paresis, and positive HITs) and central vestibular dysfunction (GEN).[5,6] HITs in unilateral vestibular nuclear infarction showed decreased gains of the VOR for the horizontal and posterior canals on both sides, but more so for the ipsilesional ones.[6]

Diagnosing CNS lymphoma is mostly challenging in the absence of pathological confirmation.[7] However, since CNS lymphoma can affect periventricular areas of the brainstem,[2,5] CNS lymphoma should be suspected in cases with a "double-panda" sign and distinctive neuro-otological abnormalities.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.3988/jcn.2022.18.1.111.

Ethics Statement

The study was conducted in accordance with the Declaration of Helsinki and its later amendments. Informed consent was obtained from the subject.

Availability of Data and Material

The datasets generated or analyzed during the study are not publicly available due to its personal information but are available from the corresponding author on reasonable request.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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